What is your diagnosis

Papules and an erythematous oozing plaque over the perianal area in an adult male: A clinical pathologic challenge

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CASE HISTORY

A 65-year-old man, known to have hypothyroidism, presented to the clinic for a few months history of progressively increasing painful lesions surrounding the anus. Physical examination was notable for a well-demarcated large erythematous oozing plaque over the perianal area (Figs. 1a and 1b). He also had a few clustered erythematous papules over the postauricular area, nasolabial folds, nose, armpits, and groin.

An excisional biopsy taken from the perianal plaque revealed diffuse dense dermal CD1a and S100 positive atypical with focal epidermotropism and mild to moderately dense accompanied by mixed inflammatory cell infiltrate (Figs. 2a – 2d). CD68 and keratin immunohistochemical stains were negative.

WHAT IS YOUR DIAGNOSIS?

Langerhans Cell Histiocytosis

DISCUSSION

The diagnosis of Langerhans Cell Histiocytosis was based on the presence of the typical histological features alongside the diffuse positivity for both CD1a and S100 markers. Langerhans cell histiocytosis (LCH) is a neoplastic disorder originating from myeloid dendritic cells that possesses heterogeneous presentations ranging from single- system to multi-system presentations [1]. Although LCH is more common in children, it can also occur in adults, often presenting as a multisystem disease (MS-LCH) with more severe, chronic, and atypical presentations. Cutaneous manifestations are common such as seborrheic-like rashes, and light-brown violaceous papules, macules, and plaques commonly affecting seborrheic and intertriginous areas [2]. Skeletal, lung, and central nervous system involvement are also common. Diabetes insipidus (DI) can also occur in cases of hypothalamic-pituitary axis involvement. Interestingly, DI has been recurrently associated with cutaneous LCH and studies have indicated that DI is the most common endocrine complication related to this condition [3].

On histopathology, LCH typically presents with a characteristic accumulation of Langerhans cells (LC), accompanied by a mixed inflammatory cell infiltrate including lymphocytes, eosinophils, and macrophages. Additionally, light microscopic examination of LCs typically reveals large mononucleated cells characterized by abundant eosinophilic cytoplasm and a distinctive 'coffee bean' or convoluted nucleus [4]. The LC differs from other histiocytes by its immunohistochemical properties, as these cells express specific markers, including CD1a, S100 protein, and langerin (CD207) [3].

Perianal histiocytosis represent the less common cutaneous manifestations of LCH with under twenty cases reported in the literature [3-5]. Perianal

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Figure 1: Erythematous plaques over the perianal area.

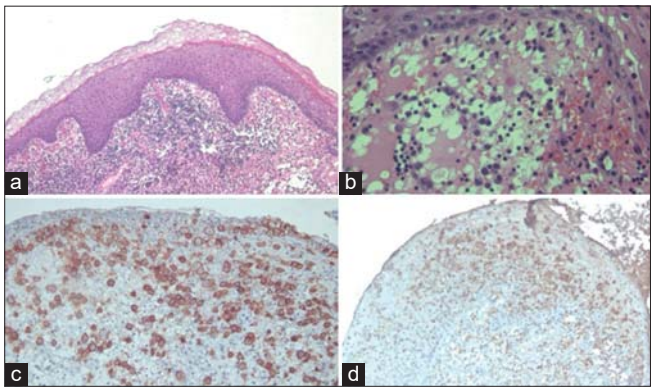

Figure 2: Photographs depicting diffuse dense dermal atypical Langerhans cells with focal epidermotropism and an accompanying mixed inflammatory cell infiltrate (hematoxylin and eosin, x20 (a) and x40 (b)). CD1a- (immunohistochemical stains, x20 (c)) and S100- (immunohistochemical stains, x20 (d)) positive atypical Langerhans cells.

histiocytosis has been described as an isolated finding or the initial symptom of multi-system LCH, pointing to the importance of considering this entity when presented with a chronic perianal lesion. Differential diagnosis includes, condyloma lata/acuminata, signet ring carcinoma, lymphoma, psoriasiform dermatitis, seborrheic dermatitis, and contact dermatitis. In our patient, the presence of papules distributed over the body raises the possibility of LCH and other infiltrative disorders and decreases the likelihood of different types of dermatitis. The diagnosis was confirmed through the typical histology and presence of confirmatory markers on immunohistochemical staining. Another diagnosis to consider in the setting of a histiocytic proliferation is indeterminate cell histiocytosis (ICH), which is typically CD1a positive. However, the diffuse S100 positivity in our case excludes the diagnosis of ICH.

For patients diagnosed with LCH, the recommended diagnostic workup involves a comprehensive approach to assess disease extent and guide treatment. In our patient, a comprehensive work-up including CBC, creatinine, liver function tests, HbA1c, TSH, urine osmolality, coagulation studies, LDH, CRP, ESR, colonoscopy, MRI scan, and a PET-CT scan was requested to complete the investigations and exclude systemic involvement. Our patient was referred to a hematologist-oncologist for further assessment and care. The patient was advised to start applying a potent topical corticosteroid once daily over the lesions until follow-up in 2 weeks, where significant improvement in the lesions was reported.

In conclusion, it is important to consider the diagnosis of LCH in the differential diagnosis of perianal lesions that have failed to resolve particularly on topical therapies, and to exclude systemic involvement after confirming a diagnosis of cutaneous LCH.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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